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Acute aortic dissection in the Marfan syndrome during the COVID-19 epidemic

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ABSTRACT

A 17-year-old boy with a recent diagnosis of COVID-19 infection was admitted with acute chest pain due to type A aortic dissection and was subsequently diagnosed with the Marfan syndrome. Literature shows an increased rate of aortic dissection during flu season. The hypothesis is that a cytokine storm triggers the dissection.

KEYWORDS Aortic aneurysm; aortic dissection; COVID-19; Marfan syndrome

Aortic dissection is the most common cause of premature death in the Marfan syndrome. Clinical diagnosis of Marfan syndrome involves multisystem findings and positive family history *(Tables 1 and 2)*. About 25% of patients with Marfan syndrome have a negative family history, and about 10% have no identifiable *FBN1* mutation. This is an index case presenting aortic complications in a young adult with the Marfan syndrome and COVID-19.

CASE PRESENTATION

A 17-year-old boy presented to the emergency department for evaluation of acute onset substernal chest pain. He was diagnosed with COVID-19 infection 2 weeks before presentation. He had a blood pressure of 130/70 mm Hg, a heart rate of 86 beats/minute, and otherwise stable vital signs. He had severe myopia and a Marfanoid appearance with a body mass index of 23.4 kg/m^2. General examination revealed a long, slender face with a high arched palate, tall body habitus, long thin fingers, pectus carinatum, pes planus, striae on the back, and a positive thumb and wrist sign. Cardiac examination revealed regular S1 and S2 with physiologic splitting. Peripheral pulses were palpable and bilaterally equal. He had no known family history of Marfan syndrome; however, his paternal grandmother had an aortic aneurysm and his paternal uncle had sudden cardiac death. Laboratory tests revealed an elevated high-sensitivity D-dimer of 526 ng/mL (reference <236); however, inflammatory markers like C-reactive protein and interleukin-6 were not checked. Computed tomography (CT) angiography of the chest revealed a fusiform aneurysm of the ascending aorta measuring 8.8 cm with a type A aortic dissection extending from the sinotubular junction to the proximal aortic arch (*Figure 1a, 1b*, Supplemental Video).

The patient underwent emergent replacement of the dissected segment with a 30 mm Gelweave with a single side branch graft. Due to a relatively less dilated aortic root and high surgical risk, aortic root replacement was not performed. The patient tolerated the procedure well without complications and was subsequently discharged. On follow-up, his genetic analysis was negative for the mutated *FBN1* gene. Postoperative CT angiography of the chest revealed a residual aortic root dilation of 4.5 cm (*Figure 1c, 1d*, Supplemental Video), and a plan was made to continue serial imaging and pursue aortic root replacement if necessary.

DISCUSSION

Cardiovascular complications of COVID-19 include pericarditis, heart failure, cardiac arrest, acute coronary syndrome, and thromboembolic events. A handful of case

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Table 1. Scoring of systemic features of the Marfan syndrome*

Systemic feature	Score
Wrist and thumb sign	3
Wrist or thumb sign	1
Pectus carinatum	2
Pectus excavatum or chest asymmetry	1
Hindfoot deformity	2
Pes planus	1
Pneumothorax	2
Dural ectasia	2
Protrusio acetabuli	2
Reduced upper segment/lower segment ratio AND increased arm span/height AND no severe scoliosis	1
Scoliosis or thoracolumbar kyphosis	1
Reduced elbow extension (≤170° with full extension)	1
Facial features (at least three of these five features: dolichocephaly, enophthalmos, down-slanting palpebral fissures, malar hypoplasia, retrognathia)	1
Skin striae	1
Myopia (>3 diopters)	1
Mitral valve prolapse	1

*Systemic score ≥ 7 is considered positive for the Marfan syndrome. The positive signs seen in the current patient are in italics.

Table 2. Diagnosis of the Marfan syndrome based on 2010 revised Ghent nosology

Family history of Marfan syndrome	No family history of Marfan syndrome
Presence of any of the following: • Aortic root dilation • Ectopia lentis • Systemic score ≥7	Aortic root dilation or dissection PLUS any of the following: • Ectopia lentis • *FBN1* pathogenic variant • Systemic score ≥7

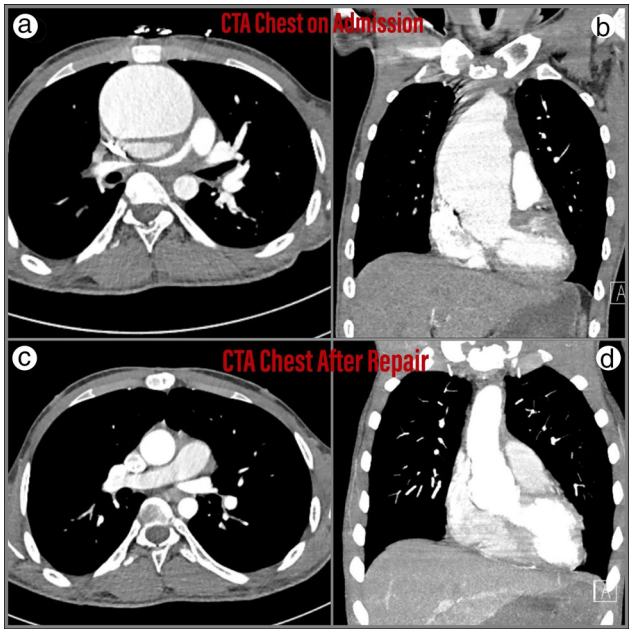

Figure 1. CT angiogram of the chest. **(a)** Anteroposterior and **(b)** coronal sections showing 8.8 cm fusiform aneurysm of the ascending aorta with type A aortic dissection. **(c)** Anteroposterior and **(d)** coronal views after aneurysm repair.

reports have linked aortic dissection to COVID-19, and most of these patients had no known predisposing genetic syndromes.[1–5] There is evidence showing an increased admission rate for aortic dissection and higher in-hospital mortality for urgent aortic repair during flu season.[6] Haidari et al demonstrated that the influenza virus could directly infect and reside in atherosclerotic arteries.[7] There is no literature suggesting a causal relation between COVID-19 and acute aortic dissection in patients with Marfan syndrome. Despite having an acute aortic dissection, our patient lacked any other COVID-related end-organ dysfunction. A study by Guo et al illustrated the detrimental effects of interleukin-1 beta in rats in promoting thoracic aortic dissection and accelerating elastic fiber fracture.[8] We hypothesize that the profound increase in interleukin-1β and other inflammatory cytokines in COVID-19 infection may be associated with worsening aneurysms in a subset of patients with preexisting syndromes causing a defective aortic wall.

In conclusion, a diagnosis of aortic aneurysm rupture or aortic dissection should be considered in patients with Marfan syndrome and COVID-19 presenting with acute chest pain. Further evidence, including animal studies, is needed to explore the direct and indirect effects of SARS-CoV2 on the aorta and its relation to the incidence of aortic dissection.

SUPPLEMENTAL MATERIAL

A CT angiogram video of the chest (left) at presentation, showing an 8.8 cm fusiform ascending aortic aneurysm with aortic dissection and (right) after aneurysm repair with a 30 mm Gelweave side branch graft.

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